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Saline agitated echocardiography in the diagnosis of cor triatriatum dexter: Case series and literature review

Short title: Saline agitated echocardiography

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INTRODUCTION

Saline agitated transthoracic or transesophageal echocardiography (SATE) is a highly effective diagnostic tool and is considered a method of choice in revealing intracardiac right-to-left shunting, e.g. through the foramen ovale (FO) or in patients with persistent left superior vena cava draining into the left atrium in case of unroofed coronary sinus. SATE may also be helpful in visualization of intracardiac flow obstruction with blood rerouting, e.g., in the cor triatriatum dexter (CTD) when prominent Eustachian valve (EV) obturates the tricuspid valve (TV), leading to pressure increase in the right atrium (RA) and right-to-left shunting *via* FO.

The main idea of SATE is to visualize the direction of microbubbles flow and reveal abnormal connections in real-time echocardiography. This is an alternative method to color

Doppler flow when multiple artifacts and lack of acoustic window cause difficulties in interpretation and decision-making process.

MATERIAL AND METHODS

The study included a retrospective analysis of two infants with hemodynamically significant CTD hospitalized in cardiac surgery department in the period of 2019-2023.

The patients were routinely diagnosed with transthoracic echocardiography, confirmed in saline agitated contrast injection and qualified for cardiac surgery.

Inclusion criteria were the following:

- a prominent EV dividing RA into two chambers with TV obturation or right-to-left shunting *via* FO;
- clinical symptoms including congestive heart failure and/or systemic desaturation;
- saline agitated injection for CTD confirmation;
- cardiac surgery with membrane excision.

Exclusion criteria were the following:

- a prominent EV in the RA without TV obstruction;
- asymptomatic course without need for SATE or cardiac surgery.

Case 1

A 6-day-old male newborn was referred for cardiac evaluation due to prenatally diagnosed complex heart defect. The child was delivered at term (40 weeks of gestation) with a body weight of 2550 g. Prenatal ultrasonography revealed agenesis of the corpus callosum and aortic atresia with large ventricular septal defect and well-developed ventricles. Genetic screening tests showed chromosomal aberrations: Jacobsen and distal trisomy 20q syndromes. On admission, the general condition was poor with symptoms of cardiac compromise and unstable vital signs: tachycardia 160/min, tachypnoea with respiratory rate 70/min, mean arterial pressure 40 mm Hg, SaO_2 85%. PGE1 and milrinone infusions were continued. Necrotic enterocolitis was observed which required conservative therapy. Paroxysmal recurrent tachyarrhythmias were registered twice with a heart rate of 250/min and were treated effectively with adenosine. In lab tests, thrombocytopenia (45 000/μl) and increased C-reactive protein (1.79 mg/dl with normal range below 0.5) were found. A blood culture was positive with *methicillin-sensitive Staphylococcus aureus* requiring antibiotics.

Transthoracic echocardiography (TTE) revealed critical heart defect with aortic atresia, extremely hypoplastic ascending aorta (2 mm) and aortic arch, large inflow ventricular septal defect, and a prominent EV dividing RA into the upstream and downstream chambers (Figure 1A–B, D; Supplementary material, *Videos S1–S4*). SATE helped to visualize and evaluate the significance of EV obstruction. The bubbles showed incomplete CTD with preserved flow into downstream chamber and right-to-left shunt *via* atrial septal defect (ASD) (Figure 1E; Supplementary material, *Video S5*). Cardiopulmonary compromise required urgent cardiac surgery with hybrid approach to avoid cross clamp circulation. Bilateral pulmonary branch bands were established and Palmaz Genesis 10 mm/19 mm stent was deployed into the arterial duct. Unfortunately, the stent displaced distally. PGE1 infusion was continued, and subsequent heart catheterization was performed with second Palmaz Genesis 10 mm/19 mm stent deployment into the proximal arterial duct, providing effective systemic flow. Inferior vena cava (IVC) angiography (Supplementary material, *Videos S6–S7*) and hemodynamic evaluation confirmed CTD with low pressure gradient between IVC and downstream chamber due to large ASD with right-to-left shunt and upstream chamber decompression. At 6 months, Glenn procedure was performed with EV resection. Postoperative hypoxia with superior vena cava syndrome required interventional left pulmonary artery Palmaz Genesis 8 mm/15 mm stent deployment. Pulmonary angiography revealed left pulmonary veins stenosis with necessity for balloon plasty.

Postoperative course was complicated with progressive cardiopulmonary compromise and hypoxia. Tracheostomy with respiratory therapy relieved the symptoms and provided SaO_2 above 80%. Following the heart team evaluation, the child was disqualified from additional interventions and at the age of 6 months was discharged to further hospice care.

Case 2

A 21-day-old female newborn was referred to cardiac surgery department with a diagnosis of CTD. In medical history, the child was delivered at term in good condition with a body weight of 3580 g. In pulse oximetry test low SaO_2 88%–92% was found. The general condition was good, and the physical examination was unremarkable except for mild central cyanosis on the mucous membrane while crying and irregular heart rate on auscultation.

The electrocardiogram and Holter study reported multiple premature atrial beats. TTE confirmed a prominent EV obstructing TV in diastole with right-to-left shunting through the FO

(Supplementary material, *Figure S1A–E*, *Videos S8–S10*). During diastole, the membrane became elongated in the shape of a windsock obstructing TV. Pulse wave Doppler showed increased velocity flow (V_{max} – 1.2 m/sec). The FO provided RA decompression with arterial desaturation. Membrane mobility predisposed to atrial premature beats. SATE supported the final diagnosis of CTD showing the severity of TV obstruction and the bubbles shunting from RA into left atrium *via* FO and helped to qualify the patient for the operation.

A cardiac surgery in cross-clamp circulation with membrane excision and FO closure relieved the symptoms. Postoperative period was uneventful. TTE showed normal TV inflow and intact atrial septum while Holter study recorded normal sinus rhythm with single premature beats. In 1 year follow up the condition was good with normal SaO_2 >95%.

DISCUSSION

In children with CTD, saline agitated echocardiography helps to visualize microbubbles outlining the intra atrial obstruction and right-to-left shunting. During the procedure, two syringes are connected by a three-way stopcock to agitate the saline by exchanging it between the 10 ml syringes. Saline is injected into a peripheral vein in the right upper extremity for FO diagnosis or into the left upper extremity for left superior vena cava diagnosis. SATE is a valuable diagnostic tool owing its minimally invasive nature. It avoids complications and radiation exposure, and provides real-time TTE assessment of intracardiac anomalies. However, patients with poor acoustic windows may have suboptimal images, risking misinterpretation [1, 2].

Understanding the embryogenesis and hemodynamics of CTD is crucial prior to SATE. During embryogenesis, the sinus venosus collects venous drainage and transforms into upstream chamber with trabeculated walls, and downstream chamber with smooth walls, right appendage and TV annulus. CTD results from persistence of the right sinus venosus valve, leading to a triatrial heart with varying degrees of partitioning and hemodynamic disturbances [3].

In very rare cases, CTD may coexist with cor triatriatum sinister, creating quadruple atria [4].

Clinical manifestations depend on the degree of TV obstruction and right-to-left shunting. Most patients, especially in adults, are asymptomatic and discovered incidentally. Thereby, due to its asymptomatic nature, complicated cannulation during surgery for other cardiac issues might occur due to fibro-muscular membrane. Besides, longer persistence of abnormal structures may

increase the risk of thrombus formation and result in pulmonary embolism [5]. In cases of long-lasting or significant TV obstruction, easy fatigability, dyspnea, hepatomegaly or even peripheral edema may be observed [6].

Screening TTE may reveal various EV morphologies. In cases of abnormal resorption of the right sinus venosus, a prominent EV may create 3 main types of CTD based on hemodynamics: free-floating EV without significant TV obstruction, incomplete CTD with persistent intra atrial communication and right-to-left shunting *via* FO, and most rarely, complete CTD with 2 distinct chambers, RV underdevelopment and right-to-left shunting *via* ASD. Surgical resection of EV may relieve the symptoms of central cyanosis or right ventricular failure.

CTD may present as an isolated anomaly or be associated with other congenital heart defects, as in patient 1. Differential diagnosis is established using echocardiography, heart catheterization with angiography, cardiac magnetic resonance imaging, or computed tomography to delineate the anatomy of the EV [6].

In conclusion, SATE is an extremely useful tool in the diagnosis of CTD with evaluation of TV obstruction, intracardiac shunts, and IVC congestion. It provides clear visualization and hemodynamic assessment which is crucial in the real-time decision making process.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/polish_heart_journal.

Article information

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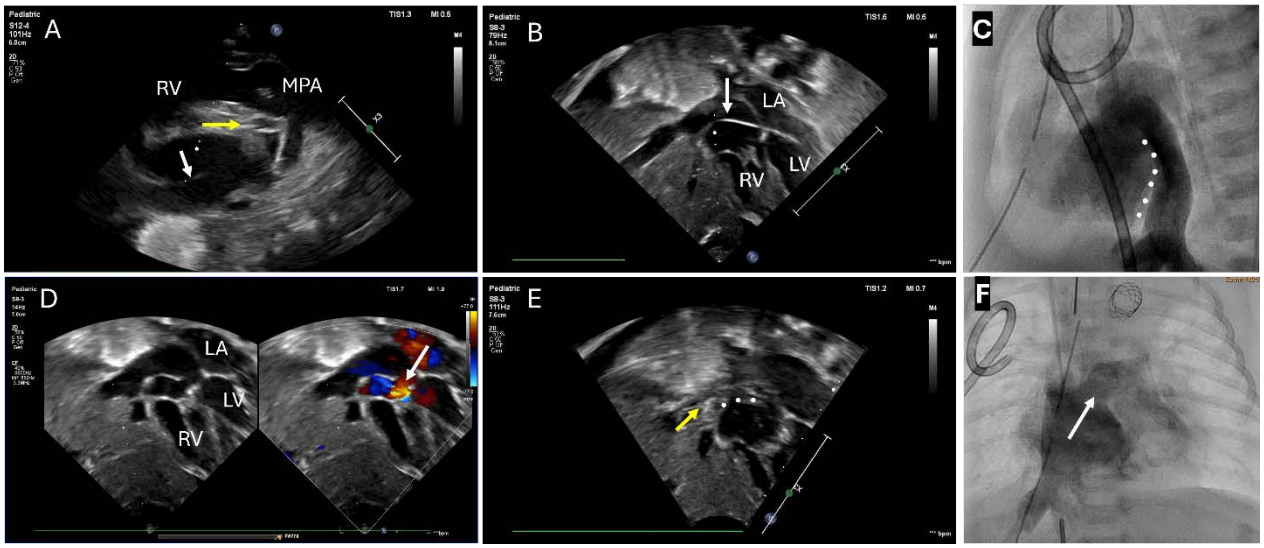

Figure 1. TTE and angiography showing cor triatriatum dexter in a patient with aortic atresia and well developed two chambers. **A.** TTE. Parasternal long axis view showing aortic atresia with hypoplastic ascending aorta (yellow arrow), wide pulmonary artery, and a prominent EV (white arrow) dividing the RA into 2 chambers. **B.** TTE. Subcostal view showing the EV dividing the right chamber into the upstream chamber with venous flow and downstream chamber with the appendage and tricuspid valve. **C.** IVC angiography in lateral view showing prominent EV (white dots) with division of the RA into 2 chambers. **D.** TTE. Subcostal 4 chamber view with color Doppler compare showing incomplete membrane of EV with small communication and laminar flow. **E.** TTE. Subcostal view showing saline agitated inflow with microbubbles (yellow arrow) in upstream chamber. White dots indicate prominent EV. **F.** IVC angiography revealing right-to-left shunt *via* atrial septal defect (white arrow)

Abbreviations: EV, Eustachian valve; IVC, inferior vena cava; LA, left atrium; LV, left ventricle; MPA, main pulmonary artery; RA, right atrium; RV, right ventricle; TTE, transthoracic echocardiography